Dear Editors,

A 45-year-old male was admitted to our outpatient for the presence of a chronic ulcer of the right lower limb associated with neutropenia lasting over 6 months. He was otherwise healthy and denied drug intake. Clinical examination showed an erythematosus ulcerated oval-shaped lesion measuring 8 cm across (Figure 1). Laboratory investigation confirmed neutropenia (3.75 × 10^9/mm^3 leucocytes with 12% neutrophils and 88% lymphocytes), Hb and platelet values being normal. Further tests included protein C, protein S, antithrombin III, lupus anticoagulant, anti-cardiolipins, ANA, ENA and ANCA: they all turned out negative as did venous Doppler ultrasound and skin swab for bacteria and fungi. Two punch biopsies were performed from the border and centre of the ulcer. Histology showed infiltration of the dermis by lymphoid elements with a large rim of cytoplasm and round-oval nuclei with dense chromatin. Similar cell population engulfed the vessels and was associated with oedema and granulomatous reaction (Figure 2). Immunohistochemistry results expressed CD2, CD3, CD8, TIA-1, CD56 and CD57, while CD20 and CD4 turned out negative. Staining for CD5 was negative, while CD7 molecule was expressed in more than 20% of the neoplastic elements. The Ki-67/MIB-1 index was below 10%. Polymerase chain reaction studies showed monoclonal TCR\(\gamma\) rearrangement. On the basis of these findings, a diagnosis of cutaneous involvement by putative T-large granular lymphocytic leukaemia (T-LGL) was made. The latter was confirmed by a peripheral blood smear that displayed circulating lymphocytes with a large rim of cytoplasm and fine azurophilic granules: on cytofluorimetry, they expressed CD16 and CD57. A trephine biopsy showed a hypocellular bone-marrow with intrasinusoidal diffusion of lymphoid elements with the same morphology and phenotype as above. Magnetic resonance and total body computed tomography scans did not detect visceral involvements. Treatment with low dose methotrexate was started. Since 3 months later the patient had not improved, Cyclosporine A (CyA) was administrated (initial dose 250 mg/die) with complete regression of the ulcerated skin lesion within a few weeks.

T-LGL is a lympho-proliferative disorder characterised by a persistent (over 6 months) increase of T-large granular lymphocytes in the peripheral blood (usually more than >2 × 10^9/l). It occurs in the sixth decade of life without sex predilection. Patients younger than 50 years are uncommon. Splenomegaly (up to 50% of cases) and chronic neutropenia are observed, while hepatomegaly and lymph node enlargement are rare (1–3). The disease is frequently related to autoimmune disorders as rheumatoid arthritis (1). Our patient had some peculiarities. First, the presentation of T-LGL in the...
form of a skin ulcer has seldom been reported (4,5). Second, the ulcer quickly improved after the start of CyA that usually represents a second line treatment (1,2). The take home message of this paper is to carefully suspect a neoplasm in case of chronic non-healing ulcers, especially if the patient is both healthy and quite young: in these cases a biopsy should always be performed.

Emi Dika, MD
Division of Dermatology
Department of Internal Medicine
Geriatric Diseases and Nephrology
Bologna
Italy
emi.dika3@unibo.it

Pier A. Fanti, MD
Division of Dermatology
Department of Internal Medicine
Geriatrics and Nephrology
Bologna
Italy

Francesco Bacci, MD
Heamatopathology Section
Sant’Orsola – Malpighi Hospital
Bologna

Alessandro Pileri, MD
Division of Dermatology
Department of Internal Medicine
Geriatrics and Nephrology
Bologna
Italy

Sabina Vaccari, MD
Division of Dermatology
Department of Internal Medicine, Geriatrics and Nephrology
Bologna
Italy

doi: 10.1111/j.1742-481X.2012.01047.x

References